



## Case report

## Adenoid cystic carcinoma of the base of the tongue: Late metastasis to the pancreas

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## ABSTRACT

Adenoid cystic carcinoma (ACC) is a relatively rare epithelial tumor of the salivary glands. We present a 64-year-old gentleman with ACC of the tongue who following resection and radiotherapy, presented 10 years later with a lung metastasis and underwent operative intervention and further radiotherapy. Five years later he presented with obstructive jaundice found to be metastatic ACC. We believe this to be the first report of an ACC metastasizing to the pancreas.

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## 1. Introduction

Adenoid cystic carcinoma (ACC) is a relatively rare epithelial tumor of the salivary glands accounting for approximately 5–10% of all salivary gland neoplasia.<sup>1</sup> It is characterized by a slow indolent growth pattern, and whilst surgical resection is the treatment of choice and usually technically feasible, local recurrence is common as a result of extensive local tissue infiltration and perineural spread.<sup>2</sup> Late metastases may also occur and have been documented most commonly in the lung but also in brain, bone, liver, thyroid, and spleen.<sup>3</sup> We present the case of a 64-year-old male patient with an ACC of the tongue who following successful resection in 1994 by means of surgery and radiotherapy, presented 10 years later with a lung metastasis with mediastinal lymphadenopathy and underwent a right lower lobectomy and lymphadenectomy followed by radiotherapy. A further five years later he presented with obstructive jaundice. Following investigation he was believed to have a cholangiocarcinoma of the distal bile duct and subsequently underwent a pancreatoduodenectomy. Histopathological examination of the specimen revealed a pancreatic neoplasm and comparison with the previous resection specimens confirmed it to be metastatic ACC. We believe this to

be the first report in the literature of an ACC metastasizing to the pancreas.

## 2. Case report

A 64-year-old male presented to our institution with a three week history of painless jaundice, generalized pruritus, pale stool and dark urine. There was no history of recent weight loss. Examination of the abdomen revealed no evidence of tenderness and no masses or organomegaly were evident. Routine biochemical investigations revealed a conjugated hyperbilirubinemia (5.1 mg/dL), and deranged liver enzymes with elevated alkaline phosphatase (227 U/L), aspartate aminotransferase (131 U/L) and alanine aminotransferase (244 U/L).

The patient's past medical history was significant for ACC of the base of the tongue diagnosed in March 1994. At that time he underwent resection of this cancer and received post-operative radiotherapy. Ten years later, in March 2004, the patient represented complaining of a lump close to the right clavicle. A computed tomography (CT) scan revealed mediastinal lymphadenopathy and a mass in the lower lobe of the right lung. He subsequently underwent a right lower lobectomy and mediastinal lymph node dissection. Pathologic investigation of the surgical specimens revealed metastatic ACC. Following this operation the patient underwent mediastinal radiotherapy.

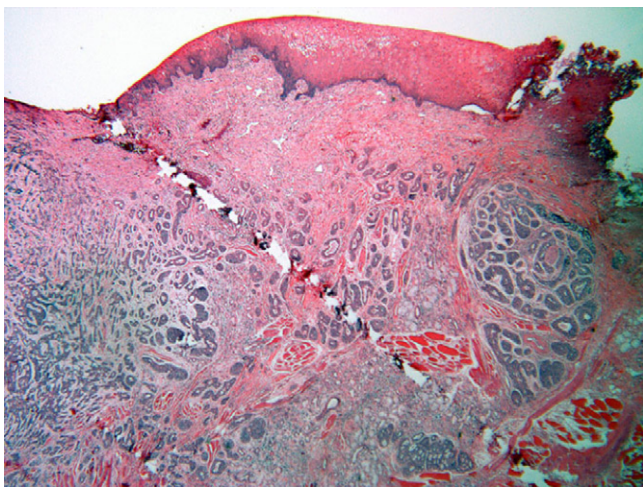
On presentation to our institution in December 2009, a CT scan of the abdomen with contrast revealed a mass in the region of the common bile duct and head of the pancreas. An endoscopic

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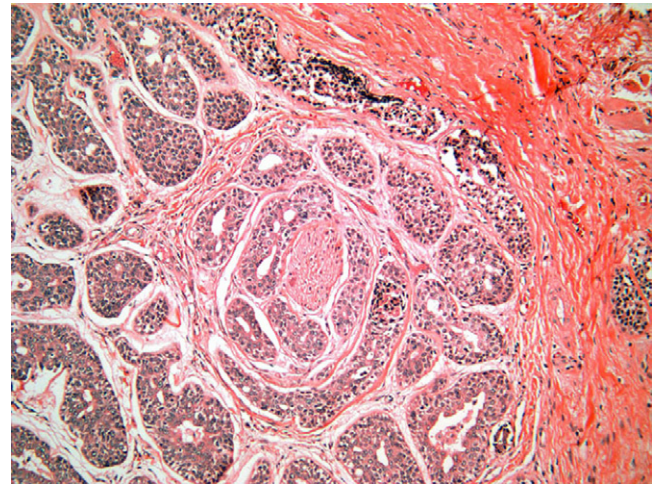
E-mail address: [falkg2@ccf.org](mailto:falkg2@ccf.org) (G.A. Falk).



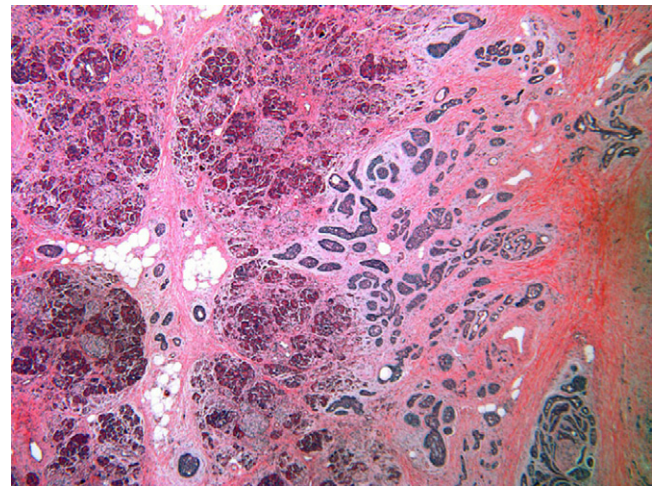
**Fig. 1.** Percutaneous transhepatic cholangiogram (PTHC) showing high-grade stricture in the distal common bile duct with a dilated proximal biliary system.



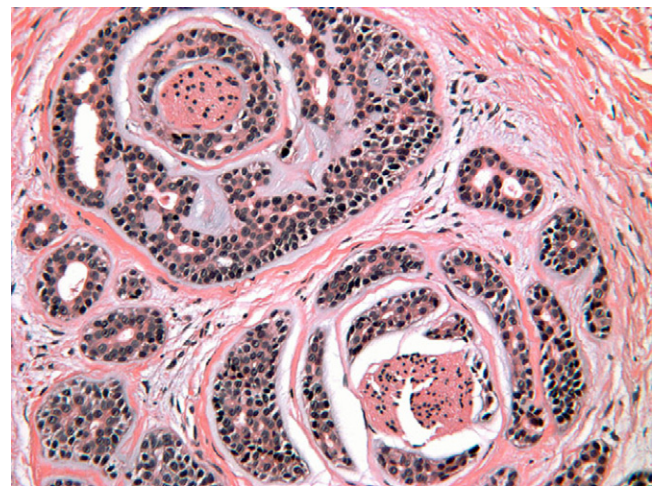
**Fig. 2.** Adenoid cystic carcinoma at the base of tongue. H&E 2 $\times$ .



**Fig. 3.** Adenoid cystic carcinoma at the base of tongue. Note neurotropic growth. H&E 20 $\times$ .



**Fig. 4.** Metastatic adenoid cystic carcinoma in pancreas. H&E 2 $\times$ .



**Fig. 5.** Metastatic adenoid cystic carcinoma in pancreas. Note neurotropic growth. H&E 20 $\times$ .

retrograde cholangio-pancreatogram (ERCP) revealed a high-grade stricture at the distal common bile duct and brushings revealed atypical cells in keeping with a cholangiocarcinoma. At ERCP it was not possible to get past the obstruction and so a percutaneous transhepatic catheter was placed for biliary decompression (Fig. 1). The decision was made to proceed with a pylorus-preserving pancreaticoduodenectomy (PPPD). At operation a mass was palpated in the head of the pancreas but there was no evidence of extra-pancreatic spread. A PPPD was performed without complication.

Histopathological examination of the PPPD specimen and review of the tumor excised from the base of the tongue and lung revealed that they were identical, indicating the pancreatic lesion represented metastatic ACC. In both sites the tumor was characterized by nests and strands of basaloid epithelial cells, with cribriform gland-like spaces (pseudoglands) and retraction artifact. Neurotropic growth characteristic of adenoid cystic carcinoma was also present in both specimens (Figs. 2–5). The tumor was excised with negative margins and at six months follow-up, the patient has no evidence of recurrence on follow-up CT.



### 3. Discussion

Adenoid cystic carcinoma (ACC) is a rare malignant neoplasm that accounts for 1–2% of all head and neck malignancies and approximately 10% of all salivary gland neoplasms.<sup>1</sup> Local recurrence of ACC is common despite aggressive surgical resection due to extensive local tissue infiltration and perineural spread. ACC is characterized by slow, indolent growth and late metastases are common.<sup>2</sup> Sites of distant metastases include lung, cerebrum, bone, liver, thyroid, and spleen.<sup>3</sup> The lungs are the most common site of distant metastases and are usually slow growing<sup>4</sup>; however, when bony metastases are present the progression is usually rapid.<sup>5</sup> Hepatic metastases of ACC are generally seen as a part of disseminated disease, with isolated hepatic metastasis being very rare.<sup>6</sup> Lymph node metastases are also rare but when present has been shown to be a negative predictor of survival.<sup>7</sup>

In the literature it appears that there is an increase in the occurrence of distant metastases in patients with a primary neoplasm located in the submandibular gland versus the parotid gland.<sup>8</sup> When the tumor recurs, neither radiotherapy or chemotherapy are unable to cure distant metastases in the lungs or at other sites but in combination with surgical resection may achieve good medium-term disease control.<sup>6</sup>

In a recent study by Opletak et al., 42% of patients with distant metastases had no evidence of loco-regional failure while the mean time between diagnosis of the primary lesion and detection of distant metastases was 59 months.<sup>8</sup> Patients with positive nodes at diagnosis lived on average 52 months less than those with negative nodes. Additionally, patients with node positive disease recurred on average 36 months earlier than those with node negative disease.

Metastatic ACC can remain dormant for many years, with pulmonary metastases being the most frequent occurrence in this setting.<sup>4</sup> Tumor stage is an important indicator of overall survival and early cancer recurrence.<sup>2,8–10</sup> The overall five-year survival rate for ACC is favorable ranging between 64% and 89%, while the 10-year survival ranges between 37% and 77%.<sup>2,9,11–13</sup>

Van der Wal et al. performed a retrospective analysis of 66 consecutive patients with ACC of the salivary glands at their institution over a 17-year period.<sup>4</sup> They found that 54% of patients had distant metastases an average of 36.8 months after diagnosis of their primary cancer. Those with lung metastasis were seen to survive approximately three years after the diagnosis of their recurrence.

### 4. Conclusion

The case presented in this paper is remarkable for a number of reasons. First of all, to our knowledge it is the first case of metastatic ACC to the pancreas reported in the literature. Secondly, the length

of time from presentation of the primary cancer to detection of the pancreatic metastases was 189 months. This case highlights the indolent nature of ACC and the importance of life-long follow-up and aggressive treatment when indicated. The awareness and use of the numerous measures available to clinicians, ranging from radiotherapy to surgical excision, can provide continued longevity to these patients.

### Conflict of interest

None.

### Funding

None.

### Ethical approval

Patient consent has been obtained.

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